Internal Biliary Fistula Secondary to Cushing's Ulcer in a Child

by Allison Ta, Suchitra Hourigan, Otto Louis-Jacques

INTRODUCTION

Internal biliary fistulas are abnormal communications between the extrahepatic biliary tree and another organ. These fistulas are rare disorders, typically affecting elderly patients as complications of biliary disease or, less commonly, peptic ulcer disease or cancer requiring surgical intervention. We present the case of a cholecystoduodenal fistula (CDF) secondary to a perforated duodenal ulcer in a pediatric patient with posterior fossa tumor. We discuss the success of medically treating this patient with proton pump inhibitors and subsequent resolution of the fistula.

Case Report

A 10 year-old female presented to the hospital with complaints of headache, vomiting, abnormal gait and upper extremity weakness for approximately eight months. She utilized ibuprofen or acetaminophen several times a week for her symptoms. Her past medical history was significant only for hypothyroidism.

A magnetic resonance imaging (MRI) of her brain showed a large posterior fossa tumor (measuring 8x4x3 cm). The patient was admitted to the pediatric intensive care unit (PICU) for stabilization and pre-operative planning. Dexamethasone was started on hospital day (HD) 1 and weaned post-operatively. On HD 4, she underwent surgical debulking of the tumor. On HD 8, she was started on nasoduodenal feeds and famotidine. The patient complained intermittently of epigastric pain, but otherwise had an uncomplicated initial postoperative course.

On HD 11, she became acutely hypotensive and her hemoglobin dropped from 10 to 5.1g/dL. Nasogastric (NG) tube lavage revealed brown colored fluid. She was resuscitated with packed red blood cells and fluids. Urgent esophagogastroduodenoscopy (EGD) discovered a large, non-bleeding duodenal ulcer. An orifice was seen medially within the ulcer bed (Figure 1). Biopsies taken at the time of EGD confirmed ulceration and ruled out Helicobacter infection. An abdominal computed tomography (CT) scan revealed air and contrast in a normal-appearing gallbladder, with a small communication between gallbladder and duodenum, consistent with a cholecystoduodenal fistula (Figure 2). Pediatric surgery was consulted and recommended medical management.

The patient was started on intravenous pantoprazole at 120 mg daily (roughly 1.6mg/kg/day), total parenteral nutrition and a NG tube was placed for gastric decompression. An EGD was repeated on hospital day 20 due to persistent melena. It showed the fistula surrounded by ulcerated tissue with a small amount of blood and bile flowing out of fistula. Attempts to place a clip on the edges of the ulcer bed were unsuccessful. Her proton pump inhibitor (PPI) was switched to continuous drip after which the bleeding resolved. Repeat EGD and abdominal CT on HD 36 showed a healed ulcer and closure of the fistula. The patient was transitioned to oral PPI and NG feeds were resumed. She had no further complications before transfer to a rehabilitation facility. At clinic follow-up more than three months later, there was no recurrence of bleeding.

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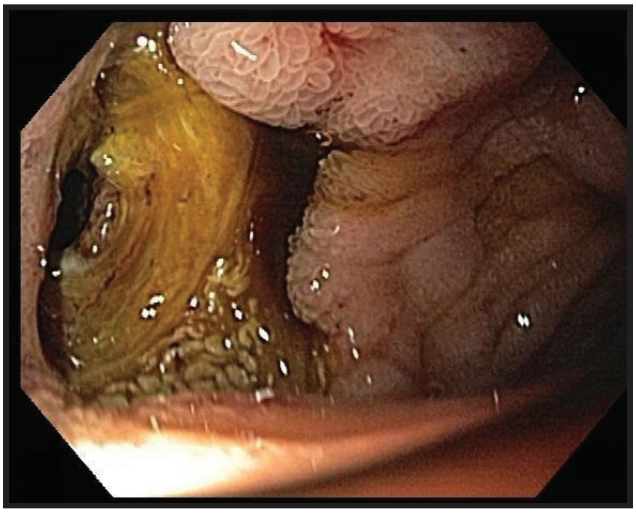

Figure 1. Perforated Internal Biliary Fistula. Initial EGD showing large ulcer and open orifice in proximal duodenum.

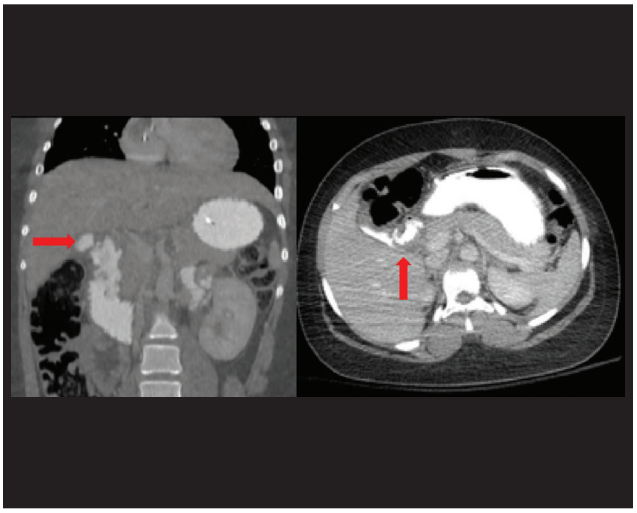

Figure 2. Abdominal CT (sagittal and coronal planes) showing contrast and air in biliary tree (arrows) consistent with a CDF.

Discussion

Internal biliary fistulas (IBF) are abnormal communications that develop between the epithelial-lined biliary tract and another organ, most commonly the gastrointestinal tract or part of the biliary tree.[1] IBF represent complications of cholelithiasis in >90% of cases.[1] Far less frequently, peptic ulcer disease, malignancy, inflammatory bowel disease or prior surgery can cause the fistula.[1] Morbidity associated with IBF is related to recurrent underlying disease, cholangitis if common bile duct is involved and/or acute gastrointestinal bleeding.[2]

The presentation of an IBF is often non-specific making its diagnosis difficult. Patients may complain of right lower quadrant pain or epigastric pain in setting of biliary disease and/or dyspepsia related to peptic ulcer disease.[3] Fever and jaundice are seen in cases of cholangitis resulting from choledochoduodenal fistulas.[2,3] Radiologic findings of pneumobilia and/or contrast in the biliary tree on plain abdominal X-ray or contrast studies (upper GI series, abdominal CT or magnetic cholangiopancreatography [MRCP]) are suggestive of the diagnosis.[4,5]

The etiology of the IBF and the stability of the patient are important in guiding management. In adults, biliary disease is commonly the cause of IBF. Definitive treatment requires cholecystectomy which allows the fistula to heal as recurrence can occur with gallstones formation.[1,5] Proton pump inhibitors are key in closure of the fistula if it is secondary to peptic ulcer disease.[3,5,6] Patients with smaller choledochoduodenal fistulas (orifice <0.5 cm) can be successfully treated medically, however surgical intervention may be necessary in case of complications from the ulcer, including uncontrolled bleeding or perforation.[5]

Given the lower incidence of gallstones and the rarity of peptic ulcer disease in children, it is not surprising that there are very few reports of IBF in children. In one case, a 6 year-old child presented with recurrent cholangitis.[2] MRCP revealed a choledochoduodenal fistula for which she underwent cholecystectomy, common bile duct excision and Roux-en-Y hepaticojejunostomy. The only previous report of a child with a choledochoduodenal fistula secondary to peptic ulcer disease is a 10 year-old male with a three month history of abdominal pain and non-bilious vomiting.[4] An EGD revealed a scarred ulcer in first part of duodenum causing obstruction which was treated surgically.

This is, to our knowledge, the first report of a pediatric patient with a CDF caused by a penetrating duodenal ulcer who was successfully treated medically. Our patient had multiple risk factors that predisposed her to developing an ulcer and subsequent bleeding including prolonged use of non-steroidal anti-inflammatory drugs (NSAIDs), the peri-operative use of systemic steroids and the stress of PICU hospitalization all in the setting of a posterior fossa tumor. An important aspect of our patient's case is the development of a

peptic ulcer in the setting of a posterior fossa tumor which has been described as a Cushing's ulcer. Harvey Cushing's initial work in 1932 described an association between intracranial tumors or injury and the development of ulceration in the upper gastrointestinal tract.[7] He theorized that these intracranial masses caused stimulation of the vagal nerve leading to increased gastrin predisposing patients to develop an ulcer in the esophagus, stomach or duodenum. This suggests prophylactic acid suppressant medication may be necessary.[7,8]

CONCLUSION

This case highlights the successful use of acid suppressants to heal a CDF caused by a perforated duodenal Cushing's ulcer, a complication rarely seen in children. In pediatrics, some patients are predisposed to develop peptic ulcer disease (NSAID use, steroids, posterior fossa tumors, to name a few) and should have prophylactic acid suppressant started. Hemorrhage from an ulcer can have high morbidity and aggressive treatment, including the use of continuous PPI infusion, should be initiated until bleeding is controlled. ■

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Answers to this month's crossword puzzle:

1 T	U	2 B	U	3 L	A	4 R	■	5 A	D	6 E	N	7 O	M	8 A
I	■	A	■	I	■	9 E	M	S	■	N	■	B	■	B
10 D	E	R	I	V	E	D	■	11 K	I	D	N	E	12 Y	S
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23 O	P	I	■	O	■	24 R	25 O	T	■	P	■	■	R	■
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32 I	33 T	34 C	H	Y	■	35 I	O	36 N	S	■	37 U	S	38 E	D
39 N	O	R	■	■	■	E	■	U	■	40 A	■	41 I	C	Y
42 I	R	O	N	■	43 I	N	F	L	A	M	E	D	■	U
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45 G	E	N	E	S	I	S	■	46 S	A	L	V	E	S	■